

MANAGEMENT OF THE CHILD WITH SPINA BIFIDA

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During the period October 1965 to September 1966 I worked at Sheffield, in England, where a particular clinical interest in the problem of spina bifida exists. The following is based largely on the teachings of that school.

Meningomyelocele is one of the most important of the congenital anomalies with an incidence of about 3 per 1000 live births in Western Europe. In the management of the individual patient all aspects must be considered simultaneously and any one specialist involved in treating one particular side of the problem must be fully aware of the other associated conditions. A formidable list of the many problems that may present are shown in the figure. Here only the major ones will be discussed.

Meningomyelocele

The main aim in surgical repair of the spina bifida is to conserve as much neural function as possible. After birth the danger of infection is considerable. Also, the neural plaque dries out, with further damage to the nerves in the sac. It has been shown in Sheffield that the earlier the lesion is closed, the better are the results as far as leg movements are concerned (Sharrard *et al.* 1963). These infants are therefore dealt with as acute emergencies and are operated within 48 hours (preferably 24 hours) after birth.

One major difficulty for the paediatric surgeon is that it is virtually impossible to assess the potential of the newborn with this defect, so that selection for surgical treatment is also nearly impossible. Only very rarely can one conclude with certainty that the baby will not survive if given normal care. In most centres virtually all cases have up till recently been submitted to surgery, but this view has now been strongly challenged and some opinions are

now in favour of some sort of selection of cases. Lorber in a recent paper has given a detailed account of the results of early surgical treatment of 524 unselected cases of meningomyelocele in Sheffield over the years 1959-63 and 1967 (Lorber 1971). In severely affected babies the overall results have been most disappointing and he concludes that those with extensive paralysis, gross hydrocephalus, severe kyphosis and those with major associated congenital defects should not be operated upon. However, Eckstein still thinks that there are probably no absolute contraindications to surgery and each case should be treated individually (Eckstein 1972). The ethical problems which are raised by the birth of a severely deformed child have been widely discussed (Illingworth and Illingworth 1965).

While waiting for surgery the baby is nursed in an incubator and the spinal defect is covered with sterile saline-soaked gauze. An X-Ray of the whole body is taken and a bacteriological swab taken from the exposed neural plaque. On the operating table, a water blanket is used to maintain the body temperature and the operation area is isolated by means of 'Vidrape' which makes it possible for the surgeon to see the leg movements in response to electrical stimulation of various areas in the plaque. The operative procedure is straightforward. The neural plaque should be isolated and preserved but all membranous tissue should be excised leaving a clean skin edge. Extensive mobilisation of skin flaps by dissecting them off the deep fascia invariably destroys most of the skin blood supply. Good results, free from complications of wound breakdown, have been obtained by Rickham using a modification of Zachary's procedure in which the skin flaps are approximated by putting sutures through the dural strip which is left attached to the

skin placing the knots inside. The approximated skin edges are sutured loosely. If the vertebral laminae are very prominent they are nibbled off (Rickham 1969).

After operation the baby is nursed prone in an incubator, with a sling under the pelvis to relieve tension on the skin sutures. Any blood loss is made good by transfusion and antibiotic cover is provided. Leakage of C.S.F. may continue for some days but is rarely a persistent complication. The two major immediate complications of meningocele are meningitis and hydrocephalus.

Meningitis is a grave neonatal emergency and on the least suspicion one must perform a ventricular tap and the C.S.F. must be examined so that the right antibiotic is given, based on sensitivities. Unfortunately, prophylactic antibiotics do not prevent meningitis; on the contrary they often lead to infections with organisms which are difficult to eradicate. It is imperative to give daily injections not only systemically but also into the cerebral ventricles, the initial antibiotic being one of the newer ones e.g.: Ampicillin, Cloxacillin, Gentamycin, Colistin, Carbenicillin, Kanamycin and Cephaloridine.

Hydrocephalus

This develops in over 80% of cases and in 100% of cases if the defect is in the thoraco-lumbar region. The commonest cause is the Arnold-Chiari malformation.

A great advance in treatment came in the early 1950s when the Spitz-Holter valve was introduced. This is a system of continuous C.S.F. drainage from the cerebral ventricles into the blood-stream via a one-way valve system. The Spitz-Holter valve is now used routinely by most paediatric surgeons, though neurosurgeons prefer modifications of it, like the Pudenz valve. Unless treatment is delayed too long, these methods allow the brain to develop normally and as a result the majority of these children, if adequately treated, are of normal intelligence. In Sheffield, routine air ventriculography is done in the first few weeks of life and this has shown that hydrocephalus is very

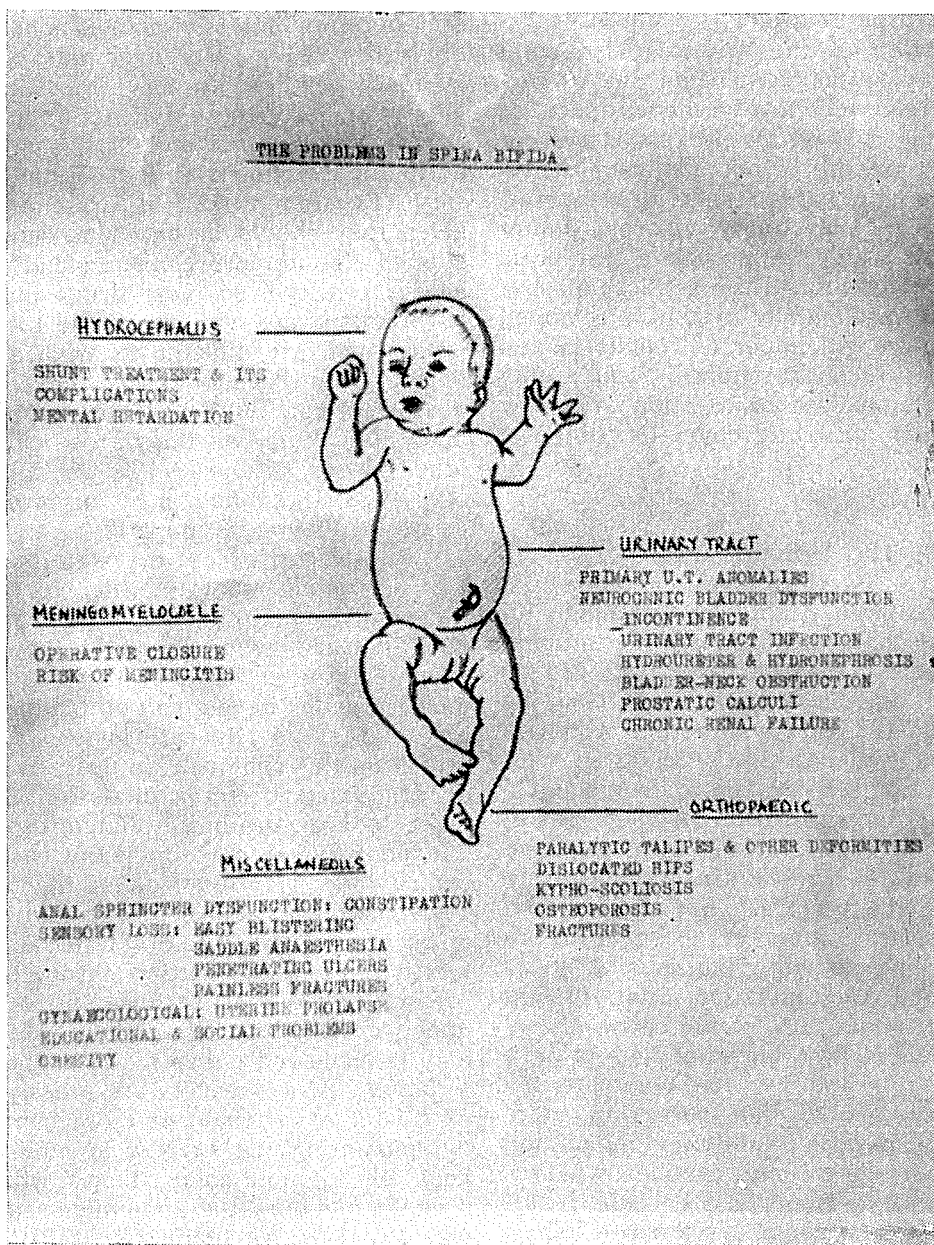
often already present then — long before the head becomes clinically enlarged.

A series of controlled therapeutic trials carried out by Dr. Lorber and his group has given remarkably precise indications for operation. If on ventriculography the pressure is less than 300 mm water and the cerebral mantle is 25 mm or more thick, then no treatment is needed. These children will have a normal-sized head and grow up to be of normal intelligence. These make up about a third of all infants with hydrocephalus. Infants with extremely severe hydrocephalus and a cerebral mantle less than 15mm thick require early shunting preferably in the first week of life. These too have a good chance of normal mental development (Lorber 1968b). The largest number of infants lies between these two groups. Here, the decision is a difficult one but no operation is usually required if the head circumference is increasing at a rate of 10mm per week or less.

The initial operation for the insertion of a ventriculo-cardiac shunt is simple. Accurate placing of the catheter up to the level of D.4 under X-Ray control is used routinely by Zachary in Sheffield and Rickham in Liverpool, but others, notably Ellison Nash do not think this is necessary because the catheter will need elective revision later on in most cases owing to the child's rapid growth.

Meningitis is an absolute contraindication to the insertion of a shunt. In such cases a Rickham reservoir is extremely useful. This is a small plastic cap which is attached to the ventricular cannula at its point of connection with the upper end of the valve. Without needling brain substance and by needling the reservoir, C.S.F. under increased pressure can be removed and antibiotics can be injected daily into the ventricles till the infection is eradicated. Then, a distal atrial catheter is incorporated into the system.

While shunt insertion carries virtually no mortality, the complication rate is alarmingly high (Tsingoglu and Forrest 1971). Blockage of the shunt at the upper end due to either the choroid plexus or growing brain substance obliterating the holes in the silastic catheter is common.



This catheter may also pull out of the ventricular cavity or it may become detached from the proximal end of the Holter valve. In this serious situation the child will have symptoms and signs of increased intracranial pressure and the Holter pump will feel flat and empty as it cannot fill up from above. The upper catheter and its connection to the valve must be 'revised'

as soon as possible. The shunt may also get blocked by organised blood clot at its lower end, either in the neck veins or in the right atrium. Occasionally, the lower end may get detached from the valve or at times may fracture. Symptoms are similar, but are slower in developing. The pump will feel stiff because one cannot empty it of its C.S.F. Treatment is not so

urgent here, but the lower catheter must be changed or 'revised'.

Infection of the shunt is also common, colonisation often being due to bacteria of relatively low virulence, such as *Staph. albus*. Low-grade pyrexia is present but blood culture is not always positive, though in some cases a spike of temperature or a rigor may follow vigorous pumping of the valve. Appropriate antibiotic treatment can control the septicaemia but unless the whole shunt system is removed the infection will linger on. Nicholas and others have recently reported good results with the immediate re-insertion of a new shunt under antibiotic cover (Nicholas *et al.* 1970).

The Urinary Tract

Spina bifida children have a high incidence of congenital urinary tract anomalies. These are however overshadowed by important complications arising from the neurogenic bladder dysfunction which is present in the majority of children with a lumbo-sacral defect. Poor detrusor muscle power, incoordination between detrusor and internal urethral sphincter and lack of normal bladder sensation leads to dribbling incontinence. There is also external urethral sphincter spasm with incomplete bladder emptying causing stasis and urinary tract infection. Scott (1970), found that about 50% of the cases assessed after their first birthday had urinary tract infection and 60% had complete urinary incontinence, this occurring more commonly in girls. Another complication is vesico-ureteric reflux leading to hydro-ureters and hydronephrosis. This is already present in about one-third of babies within a month of birth (Eckstein 1968). Hypertension and renal failure may later supervene.

The child's urinary tract should be investigated fully by means of repeated urine cultures (samples of urine are obtained by suprapubic bladder puncture), I.V. pyelograms, micturating cystic-urethrogram and, in difficult cases, cystometric pressure studies.

Prophylactic antibiotics will not red-

uce the incidence of infection and these are therefore not advisable (Zachary and Sharrard 1967). Moreover there is some evidence that the urinary tract infection may in many of these patients be localised to the bladder only and in this situation antibiotic treatment is indeed not necessary (Eckstein 1972). If antibiotics are given they should be based on sensitivity studies. The initial organism, usually *Esch. coli* is sensitive to most drugs but later re-infections are commonly due to organisms which are difficult to eradicate like *Proteus* and *Pseudomonas*. These often need treatment with parenteral injections of Gentamycin, Colistin, Carbenicillin and other newer antibiotics (Lorber and Forrby 1968). Treatment must be prolonged, usually to beyond 3 months.

Surgical treatment plays an important part in management in order to prevent progressive renal damage. As the bladder-neck is usually wide open and obstruction occurs at the external urethral sphincter, most surgeons have given up bladder-neck Y-V plasty and advocate instead a direct attack on this sphincter: in girls, by means of graduated urethral dilatation and in boys, by transurethral sphincterotomy or pudendal neurectomy. In many cases, especially in girls, diversion of the urinary tract may be needed for social reasons or because of the risks of, or actual, deterioration of renal function through back-pressure effects or because of recurrent upper urinary tract infections. Diversion may be achieved either by cutaneous ureterostomy, if the ureters are grossly dilated (Lister *et al.* 1968) or more commonly by implanting the ureters in an isolated ileal or sigmoid-colon loop, which is brought out onto the abdominal wall surface (Cook *et al.* 1968). Generally speaking, urinary incontinence in boys can be well controlled by using a penile urinal bag. If however gross hydronephrosis is present, some diversion operation is required. Such treatment is also associated with many long-term complications. The use of continuous chemotherapy is debatable, but it is usually necessary to continue with it once it has been started (Scott 1970).

Orthopaedic Problems

The lower motor-neurone lesion in spina bifida produces flaccid paralysis of both legs. The factors which determine the extent and degree of muscle paralysis are: i. — the management of the spinal defect: early closure diminishing the degree of ultimate paralysis and ii. — the level of the lesion: the legs may be completely paralysed if the highest lumbar segments are involved. Unfortunately, the large majority of children with dorso-lumbar or lumbo-sacral defects are paralysed to a variable degree. Deformities and joint dislocations result from muscle action imbalance and no amount of isolated manipulation or plaster-casting can improve the situation without reconstructive orthopaedic surgery.

Paralytic dislocation of the hips is the biggest problem and occurs in about one-half of these children. A major advance in treatment has been the development of Sharrard's operation of posterior ilio-psoas transplantation in which the still active psoas muscle is transplanted through a hole made in the wing of the ilium, fixing it to the greater trochanter of the femur (Sharrard 1971). This muscle acts as an abductor and hip stabiliser enabling the child to stand and later on to learn to mobilise himself. The operation is usually performed in the second year of life, but may be done much later. Other procedures like tenotomies, tendon transplants and osteotomies can deal with most deformities, especially those of the foot. This often means several major operations and the use of appliances supported by physiotherapy and education in walking. (Walker 1968). The use of retaining splints by irons, calipers and braces can be started in the second year of life, the object being to have the child standing at an age when the normal child does so. 'Skis' and parallel bars are necessary for walking instruction. The child is encouraged to be as mobile as possible and even a minor degree of ambulation adds greatly to the morale of the child and to that of his parents. The adoption of pessimistic attitudes in the early years will lead to com-

placent acceptance of wheel-chair life with all its drawbacks.

Educational Aspects

The intelligence range of these children without hydrocephalus is the same as that of the general child population and most of them will go to an ordinary school. The same can be said of about one-third of those with hydrocephalus, but the other two-thirds will require special schooling (Lorber 1970). In general, about 60% of children who have had treatment are of normal intelligence but because of their physical handicap many of these children have to attend special schools equipped with nursing and physiotherapy facilities. In the United Kingdom the Youth Employment Service is freely available to all school leavers and parents are encouraged to take full advantage of it. Many of these children will of course have to earn their living by mental rather than by manual work, but the more mobile child will usually have wider and better opportunities for employment.

Conclusion

In this article I have reviewed some of the most important aspects of the problems in the management of the child with spina bifida. As many specialities are involved it is most essential that treatment should be co-ordinated. Usually, liaison between the specialities is undertaken by the paediatrician, but in some centres all aspects of the child's problem are dealt with in a "combined clinic" where the paediatrician, paediatric surgeon, orthopaedic surgeon, urologist, physiotherapists and medical social worker attend at the same time. At all times, the child's parents are supported by giving them information, advice, encouragement and help.

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